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Congenital Pulmonary Atresia With Perforate Interventricular Septum in a Patient Aged Nine Years and Six Weeks

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CONGENITAL PULMONARY ATRESIA WITH PERFORATE INTERVENTRICULAR SEPTUM IN A PATIENT AGED 9 YEARS AND 6 WEEKS.

By S. B. Wolbach and Maude E. Abbott.

This case of complete congenital atresia of the pulmonary valve with perforate interventricular septum is of interest because of the age attained by the patient which was nine years and six weeks. The *clinical history* presents no remarkable features. Birth occurred in the eighth month of pregnancy and the infant weighed three pounds. Cyanosis at the age of four years was noted by the mother. The child was studied repeatedly for long periods at the Children's Hospital in Boston between the ages of four years and time of death. During this time the features were those of pulmonary stenosis. There was at all times marked cyanosis, clubbing of the fingers and toes and marked polycythaemia. The heart was markedly enlarged, at times incompetent and there was considerable bulging of the chest wall over the region of the heart. The data revealed by auscultation were consistent with the diagnosis of "pulmonary stenosis." The usual findings were fairly regular though rapid heart action with a much accentuated second sound in the pulmonary artery region. At times a systolic murmur was heard. During the later months of life acute endocarditis developed. Death occurred suddenly in the Hospital, unobserved.

The following is the report of the autopsy which was performed in the Pathological Laboratory, Children's Hospital, Boston, under serial No. A-15-3, Medical Service No. 3329, on Friday, February 10th, 1915, by Dr. S. B. Wolbach. Examination was restricted to the heart.

Gross Anatomical Diagnosis:

Complete congenital atresia of pulmonary artery; Congenital defect of interventricular septum; Aorta from both ventricles; Persistent ductus arteriosus; Acute vegetative endocarditis; Thrombosis of ductus arteriosus; Patent foramen ovale; Dilatation and hypertrophy of right ventricle with tricuspid insufficiency.

Description of Heart:

The heart is greatly enlarged and weighs 130 grams. The general shape is that of the normal heart, but the right border is rounded and it is evident that the right ventricle is enlarged. The auricles are in their normal position, the appendage of the left auricle, however, is narrow and projects as a curved process 2.5 cm. long and 0.5 cm. broad.

There is but a single arterial trunk which arises at the base of ventricles, anterior to the auricles. The tips of the right and left auricular appendages appear in their respective sides of this vessel. On opening the heart it is found that this vessel arises from both ventricles, its orifice is directly above the interventricular septum which is deficient. This deficiency corresponds to that resulting from the absence of the membranous portion of the septum, its upper border is the attachment of the anterior segment of the mitral valve; its right pillar extends to the base of the right anterior segment of the semilunar valve of the arterial trunk while its left pillar extends to the junction of the left anterior and posterior segment of the valve. The size of the interventricular opening is 2 x 1 cm. (after hardening). The arterial trunk is apparently effectively guarded by these semilunar valves, a posterior and a right and left anterior. The free edges of these segments are slightly thickened and covered with pale, grayish friable granulations.

The coronary arteries take origin behind the posterior and the left anterior segments. The size and shapes of the segments are those of a normal aorta in a heart of this size. The circumference is 8 cm. The arch corresponds to the arch of the aorta and is continuous with the descending aorta. The vessels given off are in order named: the innominate, left common carotid, a small thin walled ascending vessel and the left sub-clavian. At the location of the ductus arteriosus a large vessel takes origin, by a short trunk which divides into a right and left pulmonary artery. From this trunk, running down along the main arterial trunk is a thin walled, small calibred vessel 2 to 3 mm. in diameter. This vessel ends blindly, gradually tapering to a point in the myocardium between the posterior and left anterior segment of the valves of the main arterial trunk.

The orifice of the pulmonary trunk into the arch of the main trunk is 0.4 cm. in diameter and nearly wholly occluded by a firm grayish red clot. This clot is cylindrical in shape, 0.4 cm. long

and attached firmly over one half its circumference (organized).

The mitral and tricuspid valves are normal in appearance. The mitral measures 8.5 cm. in circumference at the attachments of the segments and the tricuspid valve measures 10 cm.

The venous orifices in the auricles are normal in number and position. The foramen ovale is patent by an oblique passage.

The endocardium is normal except for the vegetations noted on the aortic valves and a few minute vegetations on the inferior border of the opening between the two ventricles.

The myocardium is firm, of normal color and consistency. The left ventricular wall averages 1 cm. in thickness, the right 0.9 cm.

Remarks:

As has been said the interest of this case consists chiefly in the high age attained by the patient, and the fact that in spite of the unfavorable conditions present, cyanosis was not marked until after the fourth year, from which period a high degree of polycythaemia was also noted. The combination of a defect in the interventricular septum and a widely patent ductus arteriosus may supply an explanation.

Pulmonary atresia presents one of the most serious forms of congenital cardiac disease, for the complete obstruction presented to the passage of the blood from the right ventricle to the lungs implies the most extreme degree of deficient aeration possible. The duration of life is thus of course short. This is especially true of the cases with patent foramen ovale and *closed* interventricular septum which all die in early infancy and which present during life the most extreme picture of congenital cyanosis from birth, the *morbus caeruleus* of the old writers. In those cases in which the pulmonary atresia is associated with a defect of the interventricular septum at the base and a deviation to the right of the aorta so that this vessel comes to ride over both ventricles above the defect (as in the present specimen), the conditions are slightly better, and here accordingly the duration of life is considerably longer. The oldest case in the literature is one of this type recorded by Peacock, of a girl with marked cyanosis, who died at the age of 13. Here the pulmonary artery ended in an impervious funnel-shaped diaphragm formed by a fusion of the pulmonary valves, and the conus was a small canal with thin walls forming a distinct separate cavity cut off from the remainder of the right ventricle. There was a large defect of the interven-

tricular septum at the base, above which the aorta arose almost wholly from the right ventricle. The foramen ovale was closed. The ductus arteriosus was destroyed, but had probably been pervious, for a loud continuous murmur preceded and accompanied and followed the heart sounds over the whole front of the chest throughout the cardiac cycle. It is possible that here, as well as in our own case, a widely patent ductus carried the blood transmitted from the right ventricle through the septal defect into the aorta to the lungs, and thus presented conditions sufficiently favorable to the pulmonary circulation to admit of the relatively high age of 13 years being attained.

In twenty-five cases, including our own, in the literature, in six the interventricular septum was closed, and the foramen ovale was patent. In all but two of these the ductus arteriosus was also widely patent. All six were cases of extreme morbus caeruleus, and the age at death varied from three days to six months. In the remaining nineteen cases, the interventricular septum was defective; in seven of these the foramen ovale was closed, and in eleven it was also patent. The duration of life in these nineteen cases varied from thirteen days to thirteen years. In fourteen of the cases it was below five years, and in the remaining five the patients lived to the ages of six, seven, nine, eleven, and thirteen years.

The following summary of these five cases is of interest:

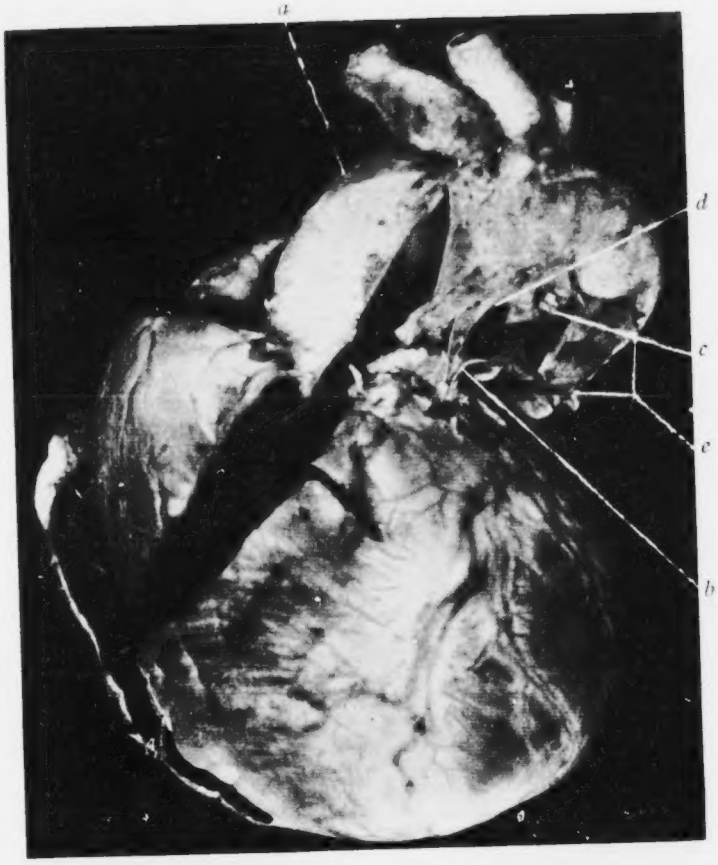
I. *Weiss* (Deut. Arch. f. klin. Med. 1875, XVI, p. 379) reports a boy of *six* with marked cyanosis and clubbing, pigeon-breast, pulmonary accentuation, but no cardiac murmurs, who died suddenly. The pulmonary artery was the size of a small pencil, and the conus was very tiny admitting a pin-head only, its endocardium was thickened and the pulmonary valve was bicuspid. The foramen ovale was patent and the interventricular septum presented a large defect at the base above which the aorta arose chiefly from the right ventricle. The ductus arteriosus was finely patent. A large anomalous branch rose from the thoracic aorta near the diaphragm and passed by a coarse anastomosis to the left lung.

II. *Moore* (Trans. Path. Soc. Lon. 1889, XLI, p. 55) records a girl aged *seven*, in whom the pulmonary artery was very small and opened out of the right ventricle through a mass of thick dense tissue which obliterated the conus. There was a large defect at the base but the foramen ovale and the ductus arteriosus were both closed.

III. *Wolbach and Abbott* (this article). From a patient aged *nine* years and *six* weeks who died suddenly. Marked cyanosis and clubbing developed at the age of four. Atresia of the pulmonary orifice, stenosis of the pulmonary conus, defect of the interventricular septum at the base, deviation to the right of the aorta, patent ductus arteriosus and closed foramen ovale.

IV. *Whipham* (Soc. Roy Med., 1910. Sec. Dis. Ch., Oct. 28, '10). A child aged *eleven* with great polycythaemia, cyanosis and clubbing, who died suddenly. The ductus arteriosus was obliterated, the foramen ovale patent, the interventricular septum defective, and the pulmonary valves were welded together so as to leave only a minute pin-hole orifice and the pulmonary artery was small.

V. *Peacock* (Trans. Path. Soc. XX, p. 61). (Cited above). The patient was a girl of *thirteen*.



CONGENITAL PULMONARY ATRESIA WITH PERFORATE INTERVENTRICULAR SEPTUM, IN A CHILD AGED NINE YEARS.

FIG. 1.—*a*. THE LARGE AORTA ARISING CHIEFLY FROM THE RIGHT VENTRICLE, *b*. RUDIMENTARY PULMONARY ARTERY ATRESIC AT ITS ORIFICE, *c*. THE PATENT DUCTUS ARTERIOSUS WITH THROMBUS, *d*. RIGHT PULMONARY ARTERY, *e*. LEFT PULMONARY ARTERY.

Reduced one-half. Dr. Wolbach's Case.

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Fig. 2.—The Left Ventricle Laid Open to Show the Aorta Riding Above the Large Defect at the Base of the Interventricular Septum, and Arising Chiefly from the Right Ventricle.